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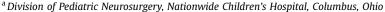
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Topical Review

Minimally Invasive Pediatric Neurosurgery

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ABSTRACT

Advances in technology have facilitated the development of minimally invasive neurosurgical options for the treatment of pediatric neurological disease. This review seeks to familiarize pediatric neurologists with some of the techniques of minimally invasive pediatric neurosurgery, focusing on treatments for hydrocephalus, arachnoid cysts, intracranial mass lesions, and craniosynostosis.

Keywords: minimally invasive, neurosurgery, pediatric, endoscopy, trans-sphenoidal, hydrocephalus, arachnoid cyst, craniosynostosis

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Historically, neurosurgical procedures have required large operative exposures to achieve the surgical goal. Such exposures can subject the patient to the risks inherent to increased brain manipulation and longer recovery times. Although many conditions are still best treated with traditional surgical approaches, improved imaging, navigation, and endoscopic technology allow more procedures to be done in a minimally invasive manner. This review seeks to familiarize pediatric neurologists with some of the techniques of minimally invasive pediatric neurosurgery, focusing on treatments for hydrocephalus, arachnoid cysts, intracranial mass lesions, and craniosynostosis.

The goal of neurosurgery is always to treat pathology with as little perturbation of normal structures as possible. In the century since Cushing formalized American neurosurgery at what is now Harvard's Brigham and Women's Hospital in Boston and Johns Hopkins in Baltimore, improvements in diagnostic imaging, surgical navigation, and endoscopic techniques have facilitated less invasive and more effective surgical procedures.

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Neurosurgical planning begins with localization of the lesion. In the beginning, the ever-important neurological examination was essentially the only tool available. Given the examination's inherent limitations, however, the surgical target could be somewhat imprecise, requiring large incisions and broad surgical exposure to ensure that the lesion would be found. The introduction of pneumoencephalography and angiography helped, but often only the secondary effects of the lesion could be visualized, not the abnormality itself. With the advent of computed tomography (CT) and magnetic resonance imaging (MRI), the lesion itself could finally be observed in increasingly precise detail. More recently, functional MRI of the gray matter and diffusion tensor imaging of the white matter have allowed even more precise planning of a minimally disruptive surgical corridor.

Once the surgical target and optimal surgical pathway have been identified, a specific operative approach must be developed for the operating room. Although every neurosurgeon should grasp the three-dimensional neuro-anatomy that facilitates this process, technological aids can be of great confirmatory utility. Computerized frameless stereotactic navigation systems allow registration of three-dimensional space of the radiographs to the three-dimensional space of the patient. Once registered, a pointer or properly prepared neurosurgical instrument can be tracked on the radiographic display showing where the instrument is and in what trajectory it is headed. This

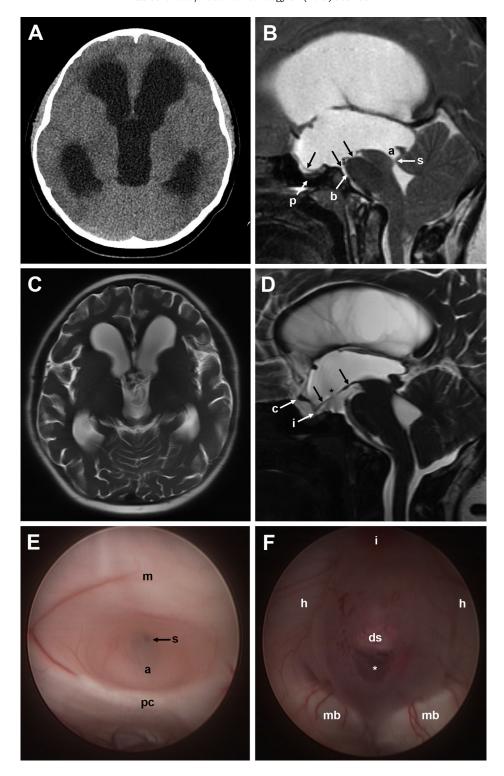


FIGURE 1.

Endoscopic third ventriculostomy (ETV). This 12-year-old girl presented with a 6 month history of syncope, headache, emesis, ataxia, urinary incontinence, and poor memory. Fundoscopy revealed papilledema and mild optic pallor. Axial computed tomography (A) revealed marked dilation of the lateral and third ventricles with transependymal absorption of cerebrospinal fluid. Sagittal T_2 magnetic resonance imaging (MRI; B) revealed aqueductal stenosis (s) and a dilated proximal aqueduct (a). The third ventricle floor (black arrows) was displaced inferiorly against the pituitary (p) and basilar artery (b). The brainstem and cerebellar tonsils were displaced inferiorly. Postoperatively, her symptoms and papilledema resolved and remained so at the 1 year follow-up. Axial single-shot fast spin-echo MRI at 4 months (C) revealed decreased ventricular size and resolution of transependymal flow. After ETV, the ventricles usually do not return to "normal" but instead establish a new baseline. Sagittal thin-cut T_2 MRI at 2 months (D) depicts the fenestration (*) with cerebrospinal fluid flow void through it. The third ventricle floor (black arrows) has returned to a normal position, and the optic chiasm (c) and pituitary infundibulum (i) are now visible. The inferior brainstem displacement and cerebellar tonsillar herniation have resolved. Endoscopic view of the posterior third ventricle floor (E) demonstrating the midbrain tegmentum (m) anteriorly, the posterior commissure (pc) posteriorly, the dilated proximal aqueduct (a), and the aqueductal stenosis (s). Endoscopic view of the anterior third ventricle floor (F) depicting the pituitary infundibulum (i) anteriorly, the hypothalamus (h) laterally, and the mammillary bodies (mb) posteriorly. The fenestration (*) lies between the dorsum sella (ds) and the basilar apex (not visible).

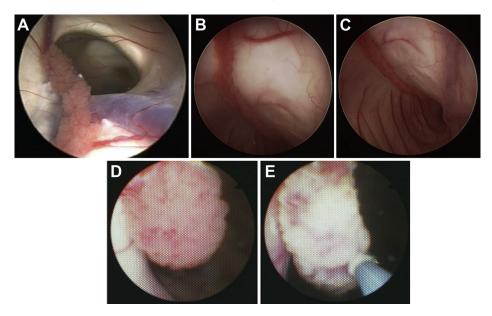


FIGURE 2. Endoscopic choroid plexus cauterization (CPC). Endoscopic images of the pink, frond-like choroid plexus as it runs in the right lateral ventricle from the foramen of Monro (A) to the ventricular body (B) to the ventricular atrium (C) and inferolateral toward the temporal horn. Images from a flexible fiber-optic endoscope during a CPC procedure depicting the choroid before (D) and after (E) coagulation.

ability can greatly facilitate precise neurosurgery, be it by open craniotomy, burr hole endoscopy, or needle/catheter puncture. Because structures can shift after opening, intraoperative ultrasound is an invaluable real-time navigation aid either by itself or integrated into the stereotactic navigation system. The ability to perform intraoperative CT and MRI can also very beneficial in certain settings.

The ability to augment one's field of vision coupled with adequate lighting is the third element in the development of minimally invasive neurosurgery. Early attempts involved loupe magnification and incandescent lights worn around the forehead. Although loupes and light emitting diode headlights certainly still have a role in neurosurgery, it was the incorporation of the surgical microscope with integrated illumination in the 1960s that allowed further

progress. Because microscopic illumination relies on an external light source providing a cone of light with its tip at the deep surgical focus, larger surgical exposures are still required. Smaller exposures would block the cone, diminish the illumination of the surgical focus, and limit what could safely be accomplished. This problem is solved with the endoscope. Endoscopic illumination allows the cone of light to be inverted with the tip at the endoscope. This allows the 3-6 mm diameter rigid or flexible endoscope to be advanced through a small aperture bringing the light to the target. Fiber-optic technology has helped channel the light to the tip of the endoscope. Coupled with lens and digital video technology, a bright high definition image is displayed for the surgeon. Specialized instruments can then be deployed around the endoscope or through working channels within it (depending on the

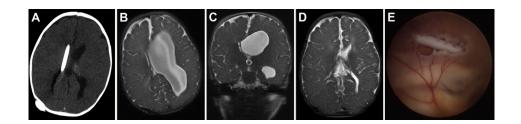


FIGURE 3.

Endoscopic septostomy. This 10-month-old girl had a right ventriculoperitoneal shunt placed at 1 month of age for hydrocephalus caused by intraventricular hemorrhage of prematurity. Because of cisternal scarring, she was not a candidate for endoscopic third ventriculostomy-choroid plexus cauterization. Axial computed tomography at 4 months revealed baseline shunted ventricle size (A). Imaging at 10 months revealed a trapped left lateral ventricle and a small right lateral ventricle (axial [B] and coronal [C] single-shot fast spin-echo magnetic resonance imaging [MRI]). An endoscopic septostomy was performed at 10 months to avoid bilateral ventriculoperitoneal shunting. Two months after septostomy (12 months old), axial single-shot fast spin-echo MRI demonstrated that the ventricles had returned to baseline size (D). Endoscopic view of the septum pellucidum from the left lateral ventricle (E). The white shunt catheter can be observed through the fenestration.

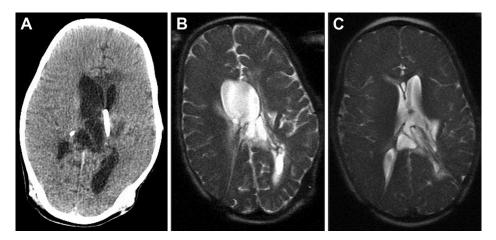


FIGURE 4.

Endoscopic fenestration of multiloculated hydrocephalus. This is a 3-year-old boy with bilateral ventriculoperitoneal shunts for multiloculated hydrocephalus from intraventricular hemorrhage of prematurity (axial computed tomography, A). He presented with steady enlargement of the right frontal horn, one of his many intraventricular cystic compartments (axial single-shot fast spin-echo magnetic resonance imaging [MRI], B). With the aid of a neuronavigation computer, an endoscope was passed into the loculated right frontal horn. Multiple cystic walls were fenestrated ultimately connecting the right frontal horn to a shunt catheter. Additionally, a path was created from one shunt to the other, which allowed later ligation of one of the shunts. After surgery the right frontal horn size normalized (axial single-shot fast spin-echo MRI, C).

circumstances of the case) to achieve the surgical goal in a minimally invasive fashion with less disruption of normal tissues, smaller incisions, and faster recovery times. Although current endoscopic instrumentation is more limited than open instrumentation, this is a focus of ongoing investigation.¹⁻⁴

Hydrocephalus

Traditionally, hydrocephalus has been treated with the implantation of a shunt system to divert cerebrospinal fluid (CSF) from the cerebral ventricles or lumbar cistern to a body cavity or into the venous system. As most practitioners know, shunts are fraught with complications. They can clog, become infected, drain too much CSF, migrate out of position, fracture, disconnect, erode through the skin or surrounding organs, and so forth.⁵ The percentage of functioning shunts decreases as the time from implantation increases with 40%-50% of all shunts failing within 2 years of implantation.^{6,7} The failure rate can be even higher in neonates. Because of this, many neurosurgeons treat hydrocephalus without shunt placement when possible. As endoscopic technology improved, techniques initially championed by pioneers like Walter Dandy were resurrected. Although Dandy was not the first to perform the endoscopic procedures for hydrocephalus, his perseverance and efforts to advance endoscopic technology were crucial to the eventual success of endoscopic neurosurgery.3

One method to treat certain types of hydrocephalus without a shunt is endoscopic third ventriculostomy (ETV). ETV results in the fenestration of the floor of the third ventricle and the underlying arachnoid membrane of Liliequist in the midline at the tuber cinereum between the mammillary bodies and the pituitary infundibulum (Fig 1). The fenestration allows flow of CSF from the third ventricle to the suprasellar cistern. It can

bypass CSF flow obstruction at the cerebral aqueduct, fourth ventricle, or fourth ventricle outlets. The lateral ventricle is accessed via a burr hole or the lateral portion of the anterior fontanel along the same transcerebral trajectory as a frontal shunt catheter. The endoscope is then passed through the foramen of Monro allowing visualization of the structures along the floor of the third ventricle. Blunt instruments are passed through working channels in the endoscope to make the fenestration. Flow through the fenestration keeps it open; a tube is not left in place.

Most frequently, the ETV is uneventful, the patient is ready for discharge the next day, and the 2.5 cm linear surgical incision behind the hairline heals well. Risks of ETV include bleeding, infection, spinal fluid leak, seizures, endocrine dysfunction, and neurological injury. Neurological structures at risk are the ones nearby, including the fornix, hypothalamus, pituitary, midbrain, cranial nerves, and basilar artery. The reported complication rate for ETV ranges from 8.4% to 13.6%. 8-10 The vast majority of these complications are transient and/or treatable, such as CSF leak, infection, seizures, hemorrhage, and transient diabetes insipidus. These series include older cases; complication rates have decreased over time. Serious complications, such as permanent neurological deficit or basilar artery injury, are exceedingly rare. Among the 604 ETV procedures reported in the three series mentioned above, for example, there were no basilar artery injuries.

The rate of ETV success in treating hydrocephalus depends on patient age, etiology of hydrocephalus, and whether the patient had a previous shunt. The chance of success can be calculated using these factors according to the ETV Success Score as elegantly elucidated by Kulkarni et al. The maximum chance of success is 90% for patients 10 years of age or greater, without a previous shunt, and whose hydrocephalus is caused by aqueductal

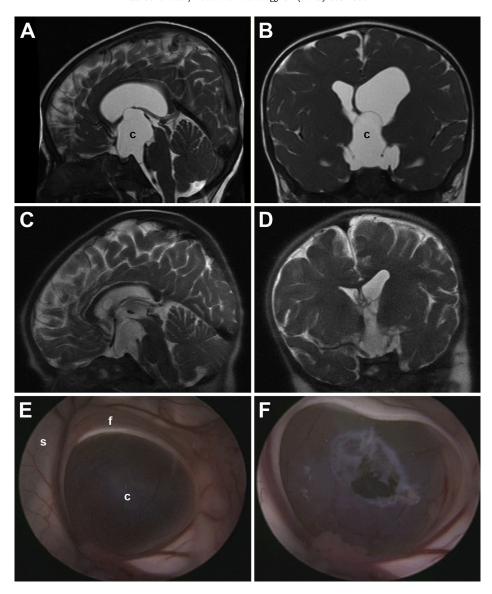


FIGURE 5.

Endoscopic fenestration of suprasellar arachnoid cyst. This 16-month-old girl presented after a syncopal episode. Sagittal (A) and coronal (B) thin-cut T_2 magnetic resonance imaging (MRI) revealed a large suprasellar arachnoid cyst (c) causing hydrocephalus of both lateral ventricles by obstructing both foramina of Monro and obliterating most of the third ventricle. Through a right frontal burr hole, the cyst was fenestrated into the ventricular system superiorly and the prepontine cistern inferiorly. Sagittal (C) and coronal (D) single-shot fast spin-echo MRI at 5 months revealed a significant reduction in the size of the cyst and the ventricular system. She had no further syncopal episodes. Endoscopic view from a different patient with almost identical pathology depicting the right foramen of Monro (E) being completely obstructed by the cyst (c), which is also stretching the overlying fornix (f). The septum pellucidum (s) serves as a midline marker, and the choroid plexus is observed posteriorly. After fenestration, the cyst collapsed away from the borders of the foramen, and the fornix is slack (F).

stenosis, tectal tumor, or other etiology not scored lower on the scale. The minimum chance for success is 0% for patients less than 1 month old whose hydrocephalus is postinfectious and who have had a previous shunt. Age plays a big role and traditionally ETV is not considered for patients less than 1 year of age. Most failures occur within 6 months of the operation. Late failures can occur, however, and caregivers must remain vigilant. 9,11

In 2000, the American pediatric neurosurgeon Benjamin Warf began to practice in Uganda for humanitarian reasons. There he found that poor infrastructure drastically increased the risk of shunting for hydrocephalus because access to care for shunt complications was limited. As much of the hydrocephalus there was congenital or perinatal, he set about finding a non-shunt method to treat hydrocephalus, knowing that ETV alone had a low success rate in this population. Some physicians suspected that the low success rate was partly due to immature CSF absorption pathways, and it had been previously shown that choroid plexus cauterization (CPC) could decrease CSF production. Warf began a trial of ETV combined with simultaneous endoscopic bilateral CPC. In 2005, he published his results. He found a 66% success rate of ETV-CPC in children less than one year of age compared with a 47% success rate of ETV alone. There was no difference in children greater than one year of

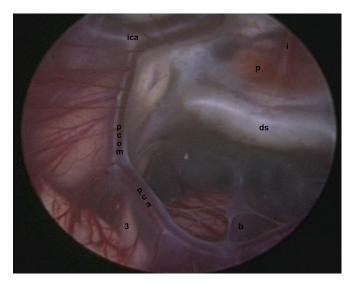


FIGURE 6

Endoscopic fenestration of suprasellar arachnoid cyst. Endoscopic view from inside the cyst depicting splayed out anatomy through the transparent inferior wall of the cyst. Visible is the pituitary (p), pituitary infundibulum (i), dorsum sella (ds), and left third nerve (3). Also apparent is the left half of the circle of Willis, including the internal carotid artery (ica), posterior communicating artery (pcom), posterior cerebral artery (pca), and basilar artery (b) traveling along the anterior surface of the pons. Stretched perforating arteries are observed originating from the circle of Willis vessels.

age. Subsequently Warf, Mugamba, and Kulkarni published the ETV-CPC Success Score.¹⁴ Factors predictive of success were patient age, etiology of hydrocephalus, and extent of CPC. Similar to ETV alone, most failures occur within 6 months of the operation. Other predictive factors in Western populations have been identified, such as width and scarring of the prepontine cistern on MRI.^{15,16}

From a technical perspective, the ETV portion is performed as previously described. Once the ETV is complete, the CPC is completed (Fig 2). Using a flexible endoscope with a monopolar coagulator in the working channel, the continuous strip of choroid plexus from the foramen of Monro to the temporal horn is cauterized. The septum pellucidum is then fenestrated, and the cauterization is completed in the contralateral lateral ventricle. The choroid plexus in the third and fourth ventricles is not cauterized. Risks are similar to ETV alone but also include venous hemorrhage from the choroid and injury to the adjacent thalamus.

Although other minimally invasive endoscopic procedures for hydrocephalus may prevent the need for a shunt in certain situations, they are more frequently used to avoid placement of additional shunts in a patient who already has one. One example is endoscopic septostomy, where a fenestration is created in the septum pellucidum to alleviate a flow obstruction from a lateral ventricle (Fig 3). Another example is endoscopic fenestration of multiloculated hydrocephalus, where membranes separating scarred portions of the ventricles are opened to allow the spaces to communicate (Fig 4).

Arachnoid cysts

Another condition that may be treatable with minimally invasive techniques is an arachnoid cyst. These CSF-filled sacs form in the subarachnoid or intraventricular spaces. Individuals with these cysts can develop hydrocephalus when the cyst blocks the flow of CSF through the ventricles (Figs 5-7). These cysts can also become symptomatic because of mass effect on the adjacent brain or skull. Most arachnoid cysts remain asymptomatic, incidental findings that require no treatment, referral, or follow-up. ^{18,19}

For the minority of arachnoid cysts that require treatment, neurosurgeons attempt to fenestrate the wall of the cyst so the fluid within can drain via normal CSF pathways. If this approach does not work or is not possible, a shunt may be necessary. Intraventricular arachnoid cysts are usually fenestrated via minimally invasive endoscopic techniques. Cysts in the subarachnoid spaces can also be fenestrated, but an open approach will allow more extensive fenestrations, lessening the chance that a shunt will be needed. Shunt avoidance is the ultimate goal because odd pseudotumor-like syndromes can be created after an arachnoid cyst is shunted. If

Intracranial mass lesions

Some brain and pituitary mass lesions, depending on their exact location and relationship to surrounding structures, can be resected using minimally invasive techniques. Using an endoscope, the neurosurgeon can access the mass through a small opening in the skull via the ventricular system (Fig 8). Other mass lesions can be accessed via a nasal trans-sphenoidal route (Fig 9). With these techniques, brain tissue disruption, incision size, postoperative pain, and patient recovery time are diminished.

Common transventricular endoscopic biopsy and/or resection targets include pineal region tumors, ²² thalamic tumors, suprasellar tumors with third ventricular extension colloid cysts, ^{23,24} and hypothalamic hamartomas. Hypothalamic hamartomas and other parenchymal lesions may be ablated using new MRI-guided stereotactic laser ablation techniques, but data regarding their complication profiles are still incomplete. ²⁶

While the trans-sphenoidal route is not new, continuously improving endoscopy and instrumentation have greatly expanded the range of what can be safely attempted through the nasal cavity (Fig 10).²⁷ In addition to lesions in or adjacent to the sella turcica, lesions of the cribriform plate, planum sphenoidale, clivus, and dens may be accessed.²⁸ Some surgeons have also used the technique to access paramedian middle and posterior fossa lesions.²⁹

Craniosynostosis

Craniosynostosis results when one or more of the cranial sutures closes prematurely. Left untreated, craniosynostosis can result in cranial deformity and, potentially, overall cranial growth restriction with resultant

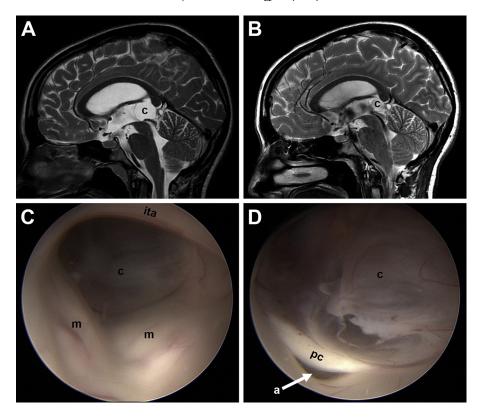


FIGURE 7.

Endoscopic fenestration of pineal region arachnoid cyst. This 16-year-old girl with Raynaud syndrome presented with several weeks of intermittent dizziness, scotomata, tiredness, posterior headache, and syncope. Sagittal T₂ magnetic resonance imaging (MRI; A) revealed a pineal region arachnoid cyst (c) compromising the proximal aperture of the cerebral aqueduct. There was associated mild lateral and third ventriculomegaly. An endoscopic fenestration was performed as was an ETV as a backup. Sagittal T₂ MRI at 4 months (B) revealed decreased size of the cyst (c) and relief of the mass effect on the aqueduct. Prominent cerebrospinal fluid flow voids are observed through the aqueduct and the ETV site. Endoscopic view of the posterior third ventricle (C) depicting the cyst (c) covering the proximal aperture of the cerebral aqueduct. Also visible is the midbrain tegmentum (m) bilaterally and the interthalamic adhesion (ita) superiorly. Endoscopic view after fenestration (D) depicting a shrunken cyst (c) that has revealed the proximal aperture of the cerebral aqueduct (a) and the posterior commissure (pc). Postoperatively, her symptoms improved but did not completely resolve. Even if she had been asymptomatic preoperatively, the imaging was concerning enough to have justified surgery.

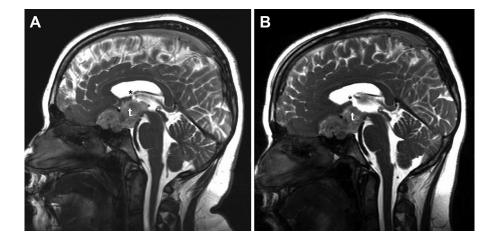


FIGURE 8.

Endoscopic tumor resection. This is a 13-year-old boy with a known hypothalamic-optic system pilocytic astrocytoma. Because of its intimate association with essential brain structures and the circle of Willis, it was not completely resectable. A On 3-month surveillance sagittal thin-cut T₂ magnetic resonance imaging (MRI; A), demonstrated tumor (t) growth into the third ventricle. Although hydrocephalus was not yet present, it would likely occur soon from occlusion of the foramina of Monro (*). To avoid bilateral ventriculoperitoneal shunt placement, we undertook a partial resection of the third ventricle tumor component. Instead of the traditional open interhemispheric transcallosal craniotomy, we performed an endoscopic resection of the tumor through a burr hole. Postoperative sagittal thin-cut T₂ MRI (B) revealed the intended result: an open third ventricle and foramina of Monro with tumor remaining attached to the hypothalamus, pituitary infundibulum, optic system, and circle of Willis. The neuro-oncologist then proceeded with medical management in an attempt to halt further tumor growth.

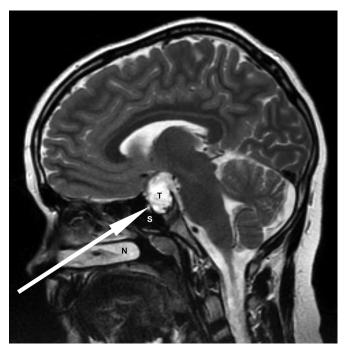


FIGURE 9. The trans-sphenoidal surgical corridor (arrow) through which many skull base lesions in the anterior cranial base, sella, clivus, and dens regions may be accessed. The most common approach passes through the nasal cavity (N), then through the sphenoid air sinus (S), then into the sella where this child harbors a tumor (T)that extends into the suprasellar region.

increased intracranial pressure. It is usually treated surgically soon after diagnosis to unlock and reshape the bones.

The traditional open surgery for craniosynostosis involves removal of at least half of the bones of the convexity, reshaping them, and reattaching them, usually in conjunction with a craniofacial plastic surgeon. It is done via a bicoronal incision across the top of the scalp from ear to ear. The surgery lasts approximately 4 hours and often necessitates blood transfusion. Postoperatively, the child is typically observed in the intensive care unit overnight followed by several days on the neurosurgical ward. Periorbital edema usually causes the eyes to swell closed then reopen before discharge. To decrease surgical risk, the operation is generally performed after 6 months of age. These children are unlikely to experience intracranial pressure sequelae of craniosynostosis before then.

The two minimally invasive options involve excision of only the fused suture to unlock the bones. Reshaping then occurs postoperatively with the assistance of either a cranial molding helmet^{30,31} or implanted custom springs.^{32,33} The bony excision is endoscope-assisted and done via one or two small incisions. The surgery lasts approximately 1 hour and blood transfusion is rarely required. Postoperatively, the child is typically observed overnight on the regular neurosurgical ward and is then ready for discharge. Usually, there is no periorbital edema. Unlike the more invasive open procedure, the postoperative reshaping adjuncts have some drawbacks. The

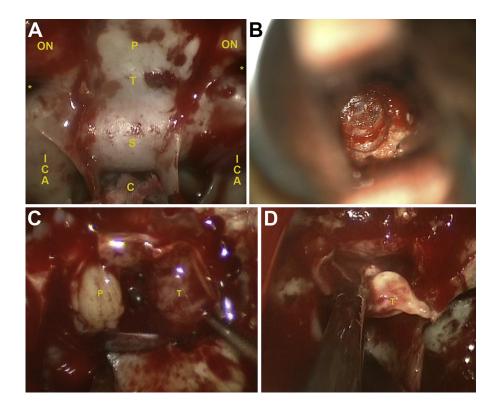


FIGURE 10

Endoscopic trans-sphenoidal skull base surgery. View of the posterior sphenoid wall from an endoscopic approach depicting a wide exposure (A). Visible are the optic nerves (ON), carotid prominences (ICA), opticocarotid recesses (*), planum sphenoidale (P), tuberculum sella (T), sella (S), and clivus (C). View of the posterior sphenoid wall and sellar dura from a microscopic approach depicting a narrow exposure limited by the width and length of the nasal speculum (B). The endoscopic approach (C) allows visual differentiation of the normal pituitary (P) from tumor (T). The endoscopic approach (D) also allows extracapsular, as opposed to piecemeal, resection of a microadenoma (T).

helmet must be worn 23 hours per day, often until the child's first birthday, and requires frequent visits to an orthotist, but not a second surgery. The springs require a second surgery for removal, but no helmet.

Which procedure to perform should be discussed by the surgeon and the family, because each technique has positive and negative attributes. Key to all options, however, is early diagnosis and referral. The age window to perform the helmet endoscopic option is approximately 2.5-3.5 months of age. The age window for the spring endoscopic option is a bit wider, but typically earlier is better for the minimally invasive options. Although minimally invasive options are more universally offered for sagittal craniosynostosis, many centers employ them for nonsagittal cases as well. Syndromic and multiple suture cases are most frequently treated with the traditional approach, but application of the minimally invasive options to this patient population is being investigated. ^{34,35}

Conclusions

Advances in technology are allowing the increasing development of minimally invasive neurosurgical options for the treatment of pediatric neurological disease. Although many lesions are still best approached using traditional methods, comprehensive centers strive to be able to offer all options. Only then can a fully informed discussion lead to the best choice for the patient by the team, of which the patient and family are a part. It is this team which allows us to achieve our ultimate goal of a healthy, happy child.

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